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Comprehensive Analysis of Lennox-Gastaut Syndrome in Europe: Treatment Patterns, Healthcare Utilisation, and Quality of Life

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Background

- Lennox-Gastaut syndrome (LGS) is a rare developmental and epileptic encephalopathy that onsets in childhood, with an estimated incidence of 0.1-0.28 per 100,000 population.[1]
- The disease is characterised by repeated and varying seizures , treatment resistance, cognitive impairments, and as a result, an impaired quality of life (QoL). [2, 3]
- There are limited real-world data exploring the population of adults diagnosed with LGS, patients' current treatment usage (including use of rescue medication), healthcare resource utilisation (HCRU) or patient QoL.

Objective

- To describe the real-world evidence of patient demographics, treatment patterns, healthcare resource utilisation and quality of life among the Lennox-Gastaut Syndrome population in Europe, stratified by different age groups, including adults.

Methods

- Data were drawn from the Adelphi Real World LGS Disease Specific Programme™ a cross-sectional survey with retrospective data collection conducted in France, Germany, Italy, Spain and the United Kingdom from July 2022 - August 2023. [4, 5] Descriptive analyses were performed.
- Neurologists and paediatric neurologists completed record forms for the next ten consecutively consulting patients with LGS. The forms collected data on patient demographics, seizure frequency and type, antiseizure medications (ASMs), rescue medications (RMs), HCRU and QoL for patients with a confirmed LGS diagnosis.
- Patient data were analysed by age groups: <6 years, 6-18 years and >18 years and by seizure types (generalised tonic-clonic (GTC), "all drop" (tonic/atonic/GTC/secondary GTC), "non-drop" (atypical absences, myoclonic, myoclonic-atonic-myoclonic-tonic-clonic, clonic, focal onset aware, focal onset impaired awareness, hyperkinetic, non-motor onset, behaviour arrest, cognitive emotional, focal with observable motor signs) seizures, experienced in the past month).

Results

Patient Demographics & Characteristics

- In total, 93 neurologists and 21 paediatric neurologists completed records for 454 patients with a physician-confirmed LGS diagnosis across five countries: France (83), Germany (78), Italy (131), Spain (118), and the UK (44).
- Mean age (Standard Deviation; SD): 16.0 years (10.8); >18 years old: 34%; Male: 65%; Mean age at first seizure: 4.0 years (2.5) (Table 1).
- In the past month, 75% had experienced drop seizures and 65% non-drop seizures. <6 years (y) and 6-18y experienced tonic seizures most commonly, and atypical seizures were most prevalent for >18y.

Table 1: Patient Demographics & Characteristics

	Overall (n=454)	<6y (n=78)	6-18y (n=221)	>18y (n=155)	All drop seizures in past month (n=342)	Non-drop seizures in past month (n=294)
Age (mean years, (SD))	16.0 (10.8)	4.1 (1.0)	12.3 (4.5)	27.2 (9.7)	15.9 (10.6)	15.8 (11.1)
Male, n (%)	297 (65)	51 (65)	150 (68)	96 (62)	229 (67)	197 (67)
Age of first seizure (mean years, (SD))	4.0 (2.5)	2.7 (1.5)	4.6 (2.2)	4.0 (3.2)	4.1 (2.6)	4.1 (2.7)
Seizures within the last month (Top 5; %)						
Any drop seizure	75	76	75	76	100	62
Any non-drop seizure	65	71	65	61	53	100
Tonic	48	60	51	37	63	43
Atypical absence	40	40	39	41	35	62
Atonic	37	37	35	39	49	35

SD, standard deviation; y, years.
For 'Drop seizures' & 'Non-drop seizures' group definitions, please see Supplementary Information.

Treatment Patterns

- Patients had a mean (SD) of 2.3 (1.5) ASM treatment regimens since diagnosis. Most common prescribed ASMs: valproate, clobazam and lamotrigine **(Figure 1).**
- RMs were prescribed to 70% of patients, consistent between age groups (70-71%). The two highest prescribed RMs were diazepam and midazolam **(Figure 2)**. The third most prescribed RM product was buccolam for all age groups apart from >18y, where clonazepam was third most common.

Overview

QUESTION What are the characteristics, treatment usage, healthcare resource utilisation and quality of life for patients with LGS across different age groups in Europe?

INVESTIGATION From the Adelphi LGS Disease Specific Programme™ [4, 5], a real-world, cross-sectional survey with retrospective data collection, conducted in France, Germany, Italy, Spain and the United Kingdom from July 2022 - August 2023

RESULTS

Sample
Records for 454 patients with a physician-confirmed diagnosis of LGS were collected.

Adult Population
34%
…of the sample were >18 years old

Most Common Seizures
3/4
…of patients across all age groups had experienced a **drop seizure** in the last month

70%
…of patients are prescribed **RMs**

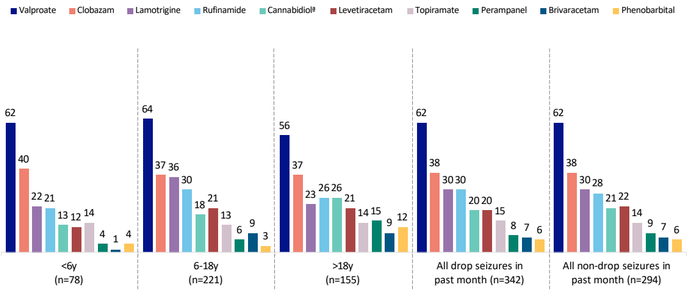
Treatment for **seizure-related complications** were the leading cause of hospitalisations in the last year, with 52% admitted for this reason

Daytime seizures were reported by physicians to have the greatest impact on patient QoL (32-49%)

47%
…of patients with LGS have **poor QoL** as reported by physicians

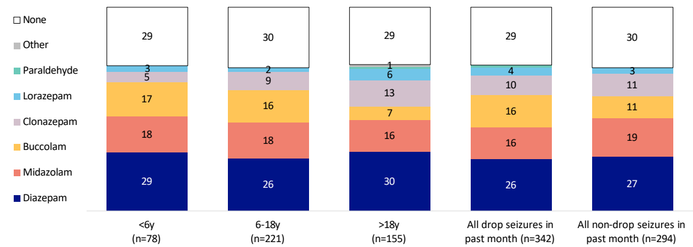
CONCLUSIONS Rescue medications are prescribed to the majority of patients with LGS, yet drop and non-drop seizures persist across the age-groups and remain the primary reason for patients being admitted to hospital within the last 12 months.

Figure 1: Currently Prescribed ASMs, Top 10 (%)



ASM, anti-seizure medications; y, years; [a]Combined cannabidiol Epidiolex and cannabidiol other than Epidiolex.
For 'Drop seizures' & 'Non-drop seizures' group definitions, please see Supplementary Information.

Figure 2: Currently Prescribed Rescue Medications (%)



RM, rescue medications; y, years.
For 'Drop seizures' & 'Non-drop seizures' group definitions, please see Supplementary Information.

Healthcare Resource Utilisation

HCRU records from the previous 12 months showed that 16% of patients had been hospitalised at least once, of whom:

- 52% had been admitted to treat a seizure-related complication;
- 92% had been admitted via the emergency room (ER);
- 19% had been admitted to the intensive care unit (ICU) **(Table 2).**

Patient QoL

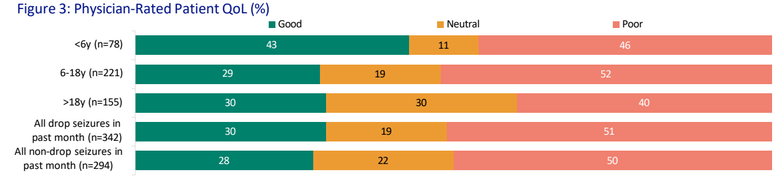
Physicians were most likely to report patients with LGS QoL as poor **(Figure 3).** Day-time seizures had the greatest impact on patients' QoL, especially in those aged <6y and >18y **(Figure 4).**

Table 2: Healthcare Resource Utilisation

	Overall LGS (n=454)	<6y (n=78)	6-18y (n=221)	>18y (=155)	All drop seizures in past month (n=342)	Non-drop seizures in past month (n=294)
Hospitalised at least once in last 12 months (%)	16	24	16	12	17	17
Admitted via ER in last 12 months (%)	92	100	90	88	92	88
Admitted to the ICU in last 12 months (%)	19	28	18	12	18	19
Reasons for hospitalisations in last 12 months (%)						
To treat a seizure-related complication	52	61	52	41	52	53
Fall/fracture following seizure	31	28	38	24	34	28
Status epilepticus	27	44	28	6	24	28
Other	17	6	21	24	20	16
Related adverse event from ASM	6	6	3	12	6	7
Hospitalisations in last 12 months (Mean (SD))	0.3 (1.0)	0.5 (0.9)	0.3 (1.2)	0.2 (0.4)	0.3 (1.1)	0.3 (0.8)
ER admissions in last 12 months (Mean (SD))	1.0 (1.6)	2.0 (2.5)	0.9 (1.3)	0.5 (1.0)	1.1 (1.7)	1.1 (1.7)

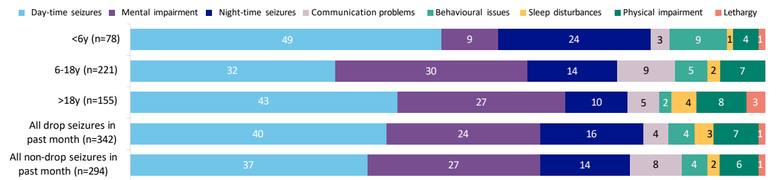
SD, standard deviation; y, years; ER, emergency room; ICU, intensive care unit; LGS, Lennox-Gastaut Syndrome.
For 'Drop seizures' & 'Non-drop seizures' group definitions, please see Supplementary Information.

Figure 3: Physician-Rated Patient QoL (%)



QoL, quality of life; y, years.
For 'Drop seizures' & 'Non-drop seizures' group definitions, please see Supplementary Information.

Figure 4: Greatest Impact on Patient QoL (%)



QoL, quality of life; y, years.
For 'Drop seizures' & 'Non-drop seizures' group definitions, please see Supplementary Information.

Conclusions

- Despite receiving multiple treatment regimens, and 70% being prescribed RMs, patients with LGS continue to experience both drop and non-drop seizures into adulthood.
- Hospitalisations were reported in 16%, almost all of whom were admitted via ER and one-fifth to the ICU.
- Around half of patients' QoL is rated as "poor" by their physicians.
- Seizures remain both the most common reason for hospitalisations as well as having the greatest impact on patient QoL.
- Findings suggest a need for new ASMs & RMs to: better control seizures, reduce HCRU and improve patient QoL.

Limitations

- The Disease Specific Programme™ (DSP) is based on a pseudo-random sample. While minimal inclusion criteria governed the selection of participating physicians, participation was influenced by their willingness to complete the survey.
- Recall bias, a common limitation of surveys, might also have affected responses. Physicians had the ability to refer to patients' records, thus minimising the possibility of recall bias.

References:

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2. Strzelczyk, A. et al. 2023. Orphanet J Rare. 18, 42
3. Gallop, K. et al. 2009. Seizure. 18(8). 554-8
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Supplementary information:

Drop seizures: tonic, atonic (drop attacks), generalised tonic-clonic, secondary generalised tonic-clonic (focal to bilateral tonic-clonic) seizures.

Non-drop seizures: atypical absence, myoclonic, myoclonic-atonic, myoclonic-tonic-clonic, clonic, focal clonic/hemiclonic, convulsive status epilepticus, non-convulsive status epilepticus, epileptic spasm, focal onset aware (simple partial), focal onset impaired awareness (complex partial), hyperkinetic, non-motor onset, focal with observable motor signs, unidentified at the time.

Disclosures

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